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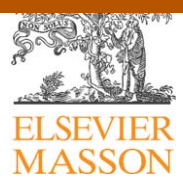

IMAGE

Late diagnosis of incomplete Cantrell's syndrome on CT scan

Diagnostic tardif d'un syndrome de Cantrell incomplet au scanner cardiaque

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A 45-year-old man with chronic myeloid leukaemia was referred for cardiac evaluation before allogeneic bone-marrow transplantation. His medical history included isolated right ventricle (RV) dilatation discovered 10 years earlier, with episodes of right heart failure induced by severe postchemotherapy anaemia.

The electrocardiogram showed first-degree atrioventricular block, complete right bundle branch block and left atrial enlargement (Fig. 1). Chest radiographs showed cardiomegaly and large bowel images in the area anterior to the heart (Fig. 2). Transthoracic echocardiography showed isolated RV enlargement (53 mm) with normal ejection fraction. Left ventricle (LV) dimension and ejection fraction were normal. The RV/LV diameter ratio was 1.2. No atrial or ventricular septal defects or pericardial abnormality was seen. Systolic pulmonary pressure estimation was 45 mmHg. Pulmonary-to-systemic blood flow ratio (Qp/Qs) was 2.1 with no visible left-to-right shunt. Right heart catheterization showed normal pulmonary artery occlusion pressure (5 mmHg), normal mean pulmonary pressure (18 mmHg) and no sign of restrictive cardiomyopathy. Oximetric study demonstrated an oxygen saturation of 66% in the superior and inferior vena cava, 84% in the right atrium, 80% in the RV and 77% in the pulmonary artery. Qp/Qs was 1.6. A computed tomography scan showed the association of transdiaphragmatic hernia, partial apical agenesis of the pericardium, and sinus venosus atrial septal defect with abnormal pulmonary venous return (Fig. 3).

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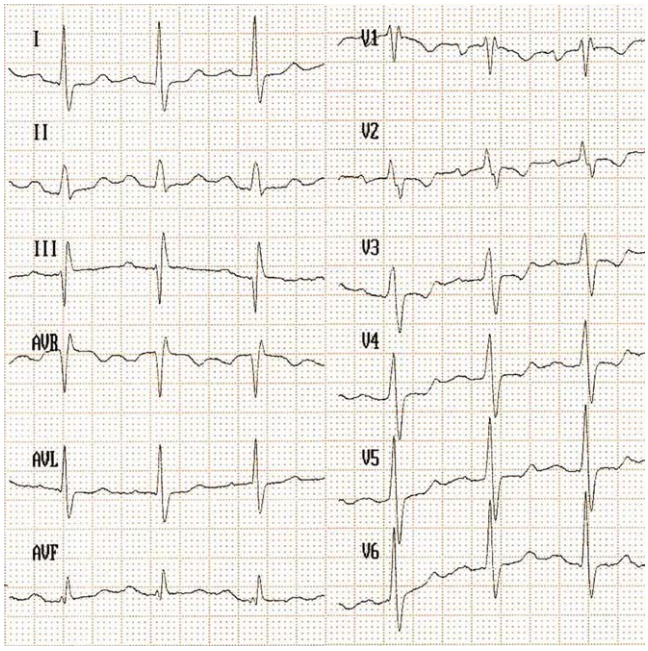

Figure 1. Twelve-lead electrocardiogram showing first-degree atrioventricular block, complete right bundle branch block and left atrial enlargement.

From these results, the diagnosis of incomplete Cantrell's syndrome in an adult patient was established. Cantrell's syndrome is a rare entity, characterized by a malformation of the lower portion of the sternum, the ventral region of the diaphragm and the mid-abdominal section, associated with cardiac anomalies. The adult form is rarely reported and is usually an incomplete form diagnosed with magnetic resonance imaging and computed tomography scanning.

Figure 2. Posteroanterior (PA) and lateral chest radiographs showing cardiomegaly and large bowel images (plain black arrows) in the area anterior to the heart.

Figure 3. (A) Axial and (B) sagittal reconstructions showing intrapericardic transdiaphragmatic hernia containing a part of the large bowel (long plain arrows) through a defect (large arrow head) of the pericardium (small arrow heads) corresponding to a partial apical agenesis of the pericardium. (C) Axial and (D) coronal reconstructions showing sinus venosus atrial septal defect (dashed arrow) with an abnormal pulmonary venous return (large arrows) in the superior vena cava (black asterisk). Enlargement of the right ventricle (panel A, white asterisk) is also observed.

Conflict of interest statement

None.